CONGESTIVE SPLENOMEGALY*
(BANTI'S SYNDROME)

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Two years ago a report[1] was presented from the Spleen Clinic of the Presbyterian Hospital outlining the results of splenectomy in Banti's syndrome. At that time we stressed the role of venous congestion or portal stasis in the production of the syndrome.

In this paper we wish to re-emphasize the same hypothesis and present additional evidence to support our contention that Banti's syndrome is not a primary splenomegaly, but rather a splenomegaly secondary to portal stasis. We have come to consider this group of splenomegalies under the general caption of "Congestive Splenomegaly," as this we feel more aptly describes the condition. The term "Congestive Splenomegaly" implies a primary congestive mechanism in the portal bed producing back pressure (splenic or portal vein hypertension). The venous stasis we believe subsequently produces a splenomegaly and certain other characteristic clinical and laboratory findings. This hypothesis is, of course, a reversal of Banti's[2,3] original idea that the disease began as a primary splenomegaly of toxic origin and later developed into cirrhosis of the liver. The conception of Banti's syndrome as a congestive splenomegaly and not a primary splenomegaly is not a new one. Warthin[4], Eppinger[5], and Larrabee[6], all considered the possibility of portal congestion producing the Banti picture. Eppinger, in fact, frequently refers to the "Stauungsmilz" in his treatise on hepatolienal fibrosis.

McMichael[7] first suggested the term "Portal Hypertension." Both he and McNee[8] agreed that the factor of increased portal pressure plus the entry of toxic substances into the portal circulation probably accounted for the development of Banti's disease. McMichael likewise intimated that a condition of portal hypertension can occur in the absence of such gross changes as are found in a hobnail liver. This latter

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statement is significant, for we have been able to demonstrate conclusively portal hypertension in many cases without cirrhosis. Portal hypertension we believe is usually secondary to one of a variety of totally unrelated diseases. Any of these conditions is capable of initiating chronic portal obstruction. Our own experience as to the types of different primary lesions which may produce congestive splenomegaly will be discussed in due course. The distended and tense venous radicals in the splenic pedicle, and the rich venous collateral often enveloping the spleen, are visible phenomena that never fail to attract anyone familiar with surgery of the spleen in congestive splenomegaly. This local vascular condition occurs regardless of the nature of the obstructive factor in any particular case. Believing that portal hypertension may be a common factor associated with a variety of disturbances producing congestive splenomegaly, members of our Clinic[9] recently have made observations on the splenic vein pressures at operation. These studies are being continued. The determinations are made at the time of operation, after mobilization of the spleen, by inserting the needle of a venous pressure apparatus into the splenic vein. Simultaneously a reading is taken of the peripheral venous pressure in one of the veins of the arm.

Splenic vein pressure readings have now been made in fourteen cases of congestive splenomegaly or Banti's syndrome, and in fifteen controls having splenic lesions of other types. In the accompanying table these figures are listed in the various sub-groupings under which we now classify our various cases.

An obstructive factor as a cause for the portal hypertension was present in 52 per cent of our early cases and no such factor was ascertainable in 48 per cent of the same group. We have found that the 48 per cent group with no obvious obstructive factor comprises chiefly the portal bed and a liver biopsy were not part of the operative our oldest cases, treated at a time when a systematic exploration of routine. However, in the past two years we have done splenectomies on twenty-four patients in addition to the previously reported thirty-one cases. With more careful inspection of the portal bed and liver biopsy on all the recent cases, and the securing of a post-mortem examination on several of the long-term cases in which an obstructive factor had previously been unknown to us, the percentage of "unknowns" is now diminishing. In these last twenty-four cases, therefore, seventeen or

TABLE I

VENOUS PRESSURES IN MILLIMETERS OF NORMAL SALINE

BANTI—LAENNEC CIRRHOSIS

	Splenic Vein	*Arm Vein*
C.M. 1.	225	12
G.M. 2.	325	85
D.P. 3.	450	125
N.A. 4.	470	145
R.B. 5.	370	30

BANTI—SCHISTOSOMIASIS MANSONI

	Splenic Vein	*Arm Vein*
P.R. 1.	250	50
A.E. 2.	335	105
G.P. 3.	500+	70
C.C. 4.	415	125
L.M. 5.	375	60

BANTI—THROMBOSIS OF SPLENIC VEIN

	Splenic Vein	*Arm Vein*
J.S. 1.	390	170

BANTI—OBSTRUCTIVE FACTOR UNDETERMINED

	Splenic Vein	*Arm Vein*
L.D. 1.	275	105 (p adrenalin)
G.K. 2.	370	50
B.S. 3.	330	55

CONTROLS

	Splenic Vein	*Arm Vein*	*Diagnosis*
F.H. 1.	190	65	Lymphosarcoma
R.B. 2.	105	80	Hemolytic Jaundice
L.L. 3.	220	205	Atyp. Hemol. Jaundice
N.B. 4.	125	130	Hemolytic Jaundice
S.S. 5.	215	40	Gaucher's
L.L. 6.	190	107	Splen. Undet. Origin
W.U. 7.	120	95	Hemolytic Jaundice
S.J. 8.	360	5 (severe shock)	Purpura
Z.G. 9.	185	75 (beginning shock)	Pancreatic Adenoma
S.L. 10.	190	210	Purpura
G.C. 11.	235	165	Splen. Undet. Origin
E.N. 12.	275	205	Atyp. Hemol. Jaundice
T.K. 13.	70	300 (10 min. later)	Purpura
M.M. 14.	140	65	Purpura
J.K. 15.	245	240	Lymphatic Leukemia

70 per cent have had a definite obstructive factor.

The number of cases of congestive splenomegaly studied by the Spleen Clinic that have had splenectomy now totals fifty-five. Sixty per cent (thirty-three cases) of the entire group have had a proven obstructive factor as a possible basis for the portal hypertension.

Under the general classification of congestive splenomegaly we have continued to use the following groupings and sub-groupings:

I. Obstructive factor known.
   A. Cirrhosis of the liver.
      a. Laennec's cirrhosis.
      b. Unclassified cirrhosis.
      c. Cirrhosis due to schistosomiasis mansoni.
   B. Thrombosis of the splenic vein.
   C. Cavernomatous transformation of the portal vein.
   D. Stenosis of the portal vein.
II. Obstructive factor undetermined.

The results of splenectomy and the long-term follow-up will now be detailed group by group under the separate disease headings (Charts I and II) as just outlined. These cases have regularly been examined and complete blood studies made at least once a year since operation.

I. Obstructive factor known.
   A. Cirrhosis of the liver.
      a. Laennec's cirrhosis:

         Fourteen cases of this type were operated upon. The immediate hospital mortality included four cases or 28 per cent. The causes of death were: shock (1); hematemesis (1); hepatic insufficiency (2).

         Seven late fatalities occurred at periods varying from five months to thirteen years. Death in all cases followed hematemesis, or cholemia with or without ascites.

         The three survivors have been well to date for periods of one and a half years, five years, and six years, respectively. These poor results are at distinct variance with those found in the succeeding groups.

      b. Unclassified cirrhosis:

         Two cases are included in this grouping; both have done exceptionally well, one for three years and one for twelve years following operation.

CONGESTIVE SPLENOMEGALY
(BANTI'S SYNDROME)
RESULTS OF SPLENECTOMY

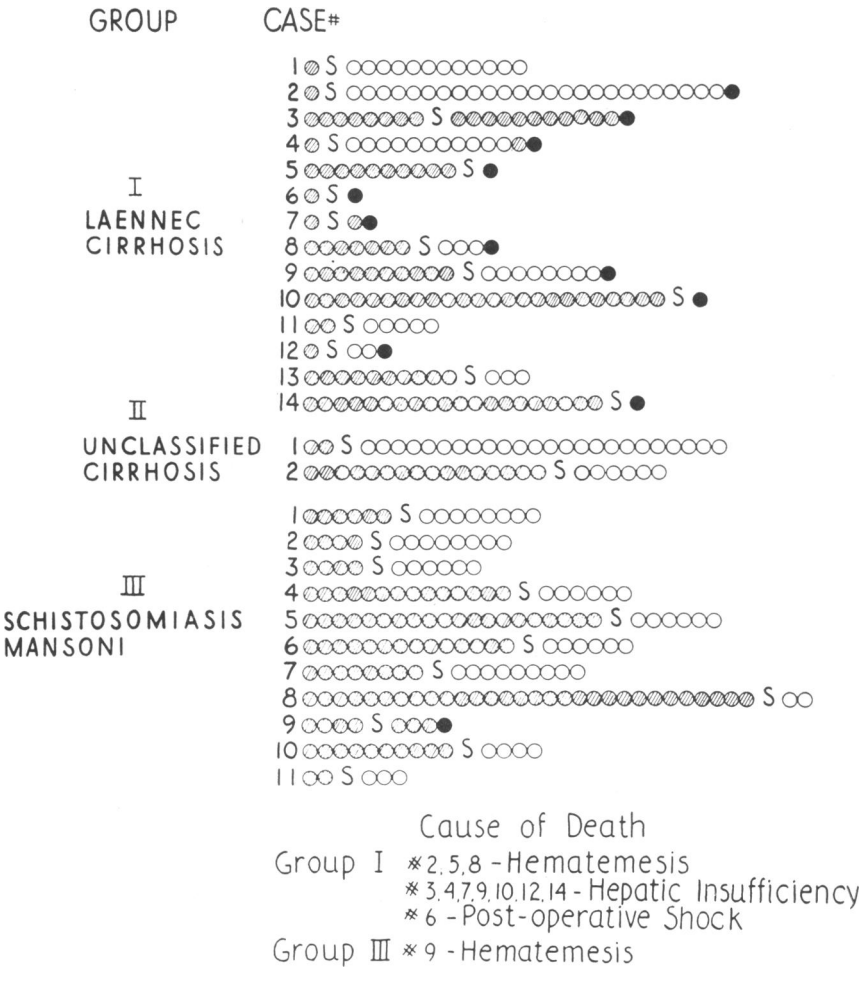

Chart I

1. **Each circle represents six months in the clinical course.**
2. **The letter S indicates time of splenectomy during the entire period patient was followed.**
3. **Clear circles denote asymptomatic periods. Shaded circles denote periods with symptoms. Black circles denote death.**

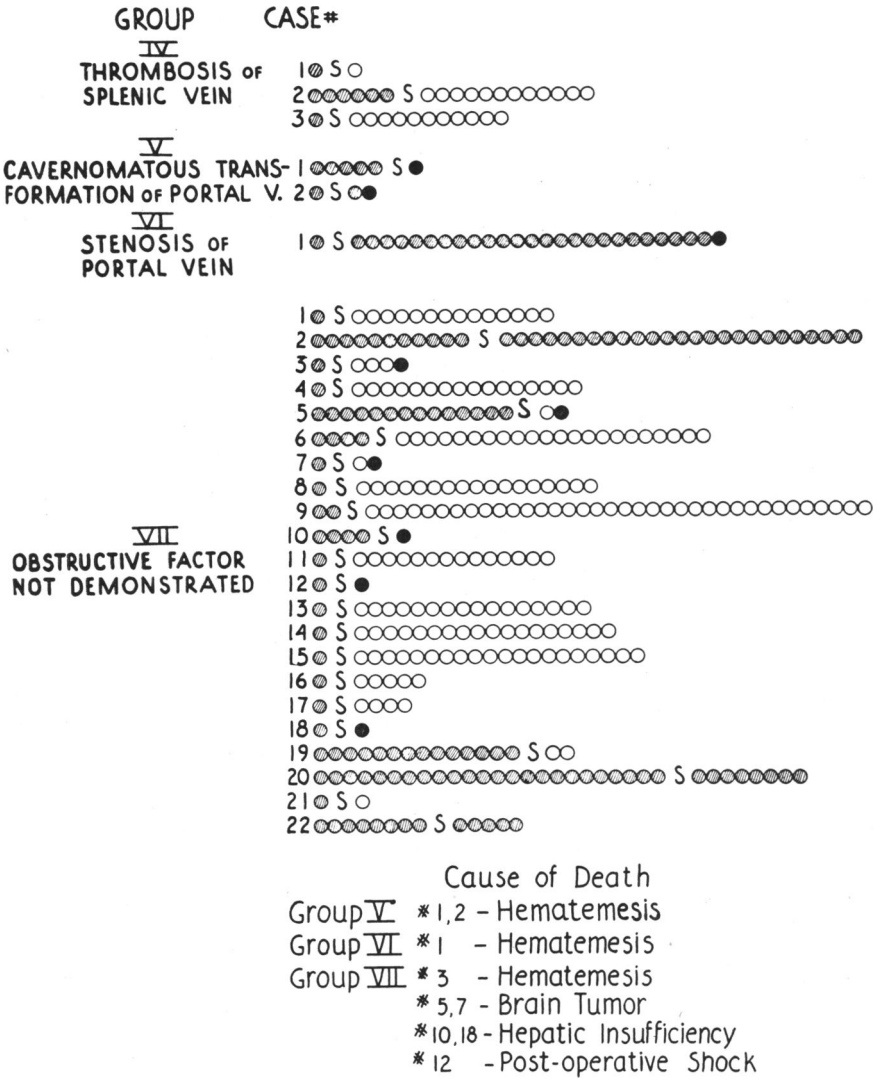

Chart II (refer to key under Chart I)

c. Cirrhosis due to schistosomiasis mansoni*:
Eleven cases of this disease with associated Banti's syndrome have had splenectomy. The mode of infection and the pathology of schistosomiasis are now well known. Faust, Jones, and Hoffman[10] have shown that the maturing phase of the parasite in the human host takes place in the hepatic,

portal and mesenteric veins. Following the dèposition of eggs in the liver, a pseudo-tubercle or fibrous nodule is formed. If the infestation is sufficiently heavy, a severe form of cirrhosis results. Coupled with this cirrhosis is a splenomegaly, and with the splenomegaly there is often a blood picture similar to that in Banti's syndrome. Oddly enough, the striking similarity of Banti's disease to the late visceral stage of schistosomiasis mansoni is one that has received scant mention. Bonelli[11] drew the clinical comparison but concluded that the Banti-like picture in late schistosomiasis was on the basis of a toxin of intestinal origin. Girges[12] with his extensive experience in Egyptian splenomegaly includes Banti's disease as a differential diagnosis of the late stage of the disease. A particularly interesting feature of our experience in these cases is that prior to operation in several of them a clinical diagnosis of Banti's disease was made and only following liver biopsy was the exact nature of the lesion ascertained. Campbell[13] in a recent publication from China has expressed the belief that most cases of Banti's disease in the Orient are really the late stage of schistosomiasis.

There has been no operative mortality in our eleven cases. The only patient who had hematemeses prior to operation remained well for a period of two and a half years after operation and then suddenly succumbed to a hematemesis. All the remaining ten patients are very well and have been followed for periods varying from two to four years.

B. Thrombosis of the splenic vein.

We have had three cases of Banti's syndrome associated with splenic vein thrombosis. All had an uneventful postoperative recovery and are well at ten months, five and a half years, and six years, respectively.

C. Cavernomatous transformation of the portal vein.

We have had two examples of this rare condition. Both cases had the typical clinical picture of Banti's syndrome with evidence of bleeding from esophageal varices. In each instance hematemesis was the cause of death following operation – in the first case, two days after operation, and in the second instance,

* A more detailed and complete report of the association of Banti's syndrome in schistosomiasis and the results of splenectomy in this disease, is in press.

nine months after operation. Of particular interest to us is the fact that the liver was normal in each of these cases, proven by autopsy; hence it is obvious that hematemesis is not necessarily the sequel of cirrhosis but can be a fatal issue in the presence of obstructive lesions other than cirrhosis. One of these cases was particularly instructive to us because prior to operation all the liver function tests gave normal values; at operation the liver grossly appeared normal and the liver biopsy likewise showed normal liver structure. We therefore classified this as one of the cases in which the obstructive factor was undetermined. It was only when the patient died nine months later that autopsy demonstrated the obstructive factor at the beginning of the portal vein. Klemperer[14] reported one such case and was able to collect twenty-three similar examples in the literature. His patient, as ours, also had undergone a splenectomy for supposed Banti's disease several years prior to death.

D. Stenosis of the portal vein.

This is a recent addition to our list of possible lesions causing portal congestion. We have had one patient with this unusual condition. At operation no obstructive factor was demonstrated. The liver biopsy was normal. Following operation the patient had six hematemeses over a period of thirteen years, with finally a fatal hemorrhage. At necropsy there was no cirrhosis, but a stenosis of the portal vein was present near its origin.

II. Obstructive factor, undetermined.

Our last series in the congestive splenomegaly group includes twenty-two cases in which the obstructive factor could not be demonstrated at operation. Three died in the postoperative period, a mortality of 13.6 per cent. The cause of death included shock (1); hepatic insufficiency (2).

Only one late death occurred, and this, two years and two months after operation, of hematemesis.

Two died several years after operation of an unrelated disease.

Of the remaining sixteen, thirteen have been entirely well for periods varying from two months to eighteen years. However, there are three patients still living who have had a miserable course punctuated by repeated, severe hematemesis over periods of three years, four years, and two and a half years, respectively.

SUMMARY

In a group of fifty-five cases, similar characteristics were apparent in all, to warrant a diagnosis of congestive splenomegaly, or so-called Banti's syndrome. Evidence has been presented to show that portal hypertension exists as a common factor in a variety of different clinical entities. Each of these various diseases is capable of producing chronic portal stasis.

The indications for splenectomy in congestive splenomegaly and the long term prognosis are dependent on the nature and severity of the obstructive factor. Obviously, splenectomy is contraindicated in cases of progressive decompensated liver disease. The late results in our cases with Laennec's cirrhosis, portal vein occlusion or stenosis, have been extremely poor. The results in other forms of cirrhosis, splenic vein thrombosis, and the group in which the obstructive factor was undetermined, have been most gratifying. Hematemesis as a postoperative symptom is usually a grave prognostic omen.

REFERENCES

1. Rousselot, L. M. The role of congestion (portal hypertension) in so-called Banti's syndrome, *J. A. M. A.*, 1936, *107:* 1788.
2. Banti, G. Splenomegalie mit Lebercirrhose, *Beitr. z. path. Anat.*, 1898, *24:* 21.
3. Banti, G. Über Morbus Banti, *Folia haemat.*, 1910, *10:* 33.
4. Warthin, A. S. Relation of thrombophlebitis of the portal and splenic veins to splenic anemia and Banti's disease, *Internat. Clin.*, 1910, Ser. 20, *4:* 189.
5. Eppinger, H. *Die hepato-linealen Erkrankungen.* Berlin, Springer, 1920, pp. 384-493.
6. Larrabee, R. C. Chronic congestive splenomegaly and its relationship to Banti's disease, *Am. J. M. Sc.*, 1934, *188:* 745.
7. McMichael, J. Pathology of hepatolienal fibrosis, *J. Path. & Bact.*, 1934, *39:* 481.
8. McNee, J. W. The spleen; its structure, function and diseases (Lettsonian lecture), *Lancet*, 1931, *1:* 951; 1009; 1063.
9. Thompson, W. P., Caughey, J. L., Whipple, A. O. and Rousselot, L. M. Splenic vein pressure in congestive splenomegaly (Banti's syndrome), *J. Clin. Investigation*, 1937, *16:* 571.
10. Faust, E. C., Jones, C. A. and Hoffman, W. A. Studies on schistosomiasis mansoni in Puerto Rico; the mammalian phase of the life cycle, *Puerto Rico J. Pub. Health & Trop. Med.*, 1934, *10:* 133.
11. Bonelli, P. Analogia entre la Schistosomiasis mansoni y la enfermedad de Banti o anemia esplénica, *Bol. Asoc. méd. de Puerto Rico*, 1931, *23:* 251.
12. Girges, R. *Schistosomiasis (bilharziasis).* London, Bale, 1934.
13. Campbell, H. E. Splenomegaly in the Foochow area, with special reference to schistosomiasis, and its relationship to cryptogenetic splenomegaly (Banti's disease), *Chinese M. J.*, 1936, *50:* 1561.
14. Klemperer, P. Cavernous transformation of the portal vein; its relation to Banti's disease, *Arch. Path.*, 1928, *6:* 353.